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Title: Congenital Extrahepatic Portosystemic Shunt: Abernethy Malformation Type 2

Running head: Abernethy Malformation Type 2

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INTRODUCTION

Abernethy malformation (AM) with congenital anomaly was defined by John Abernethy in 1973. This malformation is characterized by shunting and is between the portal vein (PV) and the systemic circulation (1, 2). AM is frequently associated with other rare congenital anomalies, including the extrahepatic portocaval shunt, heterotaxy, biliary atresia and liver nodules (3,4). There are two types of AM which have been classified into two types based on the pattern of anastomosis between the systemic circulation and PV and the presence an intrahepatic portal venous supply. AM Type I portosystemic shunts is characterized by complete shunting and absence of a PV. AM type II is characterized by partial shunting with a small grade of PV flow to the liver (5). Assessment of the vascular anatomy and liver by new abdominal imaging technological in AM aids planning, so that the patient can receive appropriate treatment. Alternative treatment mode non-surgical endovascular treatment, if this treatment fails, the liver transplantation may be considered (5). In this study, we present the case of a male who was incidentally diagnosed with AM of type II.

CASE PRESENTATION

A 68-year-old male patient presented to the gastroenterology department with complaints of abdominal pain and nausea. The patient had no significant personal or family medical history. No pathology was found except the right upper quadrant sensitivity on physical examination. The patient underwent total gastrectomy for gastric adenocarcinoma 3 months ago. Laboratory tests showed hemoglobin 13 g/dL, direct bilirubin 1.24 µmol/L, indirect bilirubin 3.70 µmol/L, lactate dehydrogenase 322 U/L, alanine aminotransferase 21 U/L, aspartate aminotransferase 40 U/L, gamma-glutamyl transpeptidase 32 U/L, alkaline phosphatase 116 U/L, prothrombin time 39.0 sec. Serologic tests for hepatitis B and C viruses negative results. Abdominal ultrasonography showed the presence of an anechoic tubular structure approximately 13 mm in diameter in the liver with no current Doppler signal. Computed tomography (CT) axial sections showed superior contrast enhancement in the right lobe of the liver, suggesting a 5 mm diameter hemangioma. PV diameter was measured as 5 mm. PV superior mesenteric vein (SMV) and splenic ven junction left renal vein portocaval shunt. (Figure A, B) Given these radiological findings, the hypoplastic PV with a portocaval shunt (AM of type II), The patient continued conservative treatment and after 1-month abdominal pain and nausea symptom was mild. At 3-month follow-up, he was no change on ultrasonography. Firstly the patient will consider the alternative treatment mode non-surgical endovascular treatment. If this treatment fails, the liver transplantation may be considered. (Informed consent is taken from the patient before writing this report)

DISCUSSION

Congenital anomalies and vascular shunt diseases are seen together (6). AM can be anatomically classified with radiological imaging techniques. AM type II portosystemic shunt is characterized by the presence of a patent intrahepatic portal venous supply and a partial shunt (7). Type I can be further subclassified into type Ia and type Ib. Type Ia, separate drainage of the SMV and splenic vein into systemic veins, type Ib where SMV and splenic vein join to form a short extrahepatic portal vein which drains into a systemic vein. This patient had a type II AM with a side-to-side portocaval shunt between the left renal vein and splenic vein. Congenital vascular malformations are frequently associated with congenital anomalies. Other anomalies have also been reported in patients with AM which include chromosomal anomalies such as Down syndrome and structural anomalies of the cardiac defects, biliary atresia, polysplenia, situs inversus (8,9,10). Hepatic shunt frequently can also present with hypoglycemia. This might be the effect of defective uptake of glucose and defect of insulin secretion due to reduced hepatic degradation of the normal quantity of secreted insulin (11,12). A diagnosis of AM can now be made by noninvasive abdominal imaging technologies, such as USG, CT, and Magnetic Resonance Imaging (MRI) (13). The imaging findings in patients with AM with hepatocellular carcinoma do not appear to be typical, that is hypervascularity on the arterial phase images with washout on delayed phase (14). Patients who do not have typical findings of a benign lesion, i.e. lack arterial enhancement or arterial enhancement without washout, should be closely followed up. Two groups according to the type of shunt those should be offered shunt closure either interventional embolization or surgical whereas those with type 1 shunts should be liver transplanted (15,16).

CONCLUSION

AM is a rare congenital vascular malformation that can be diagnosed to abdominal imaging technological (USG, CT, MRI). In this case, we present the case of a male who was incidentally diagnosed with AM of type II. Endovascular treatment of Abernethy malformation type 2 should be the first-line treatment if not successful liver transplantation should be considered

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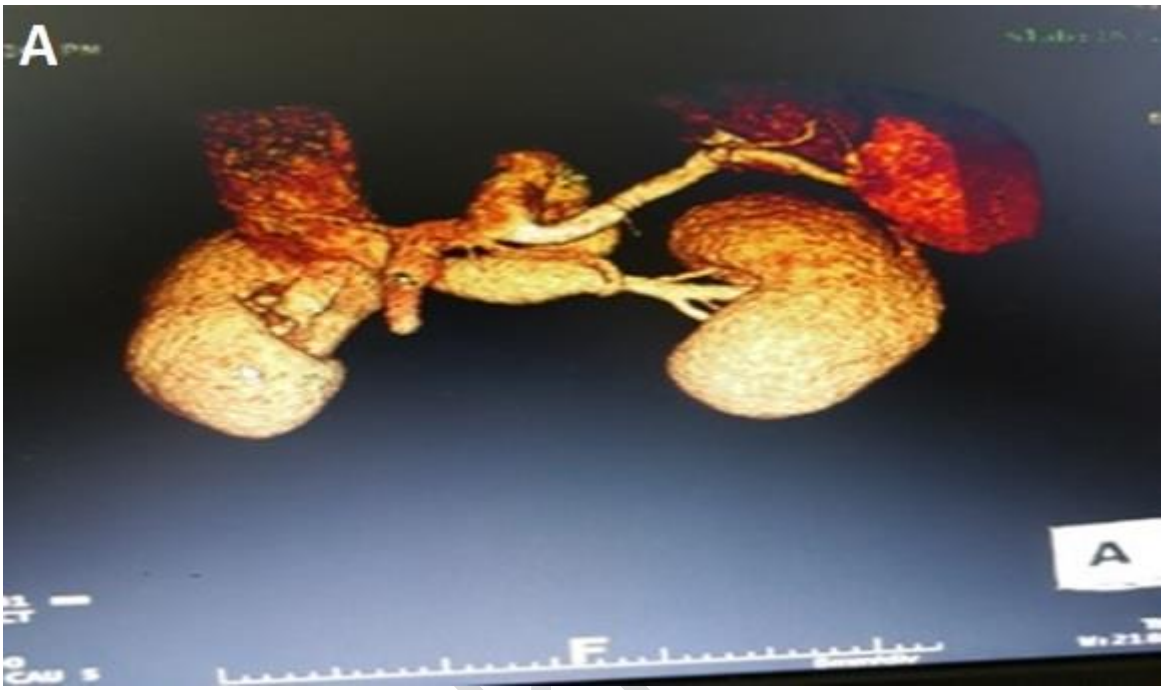
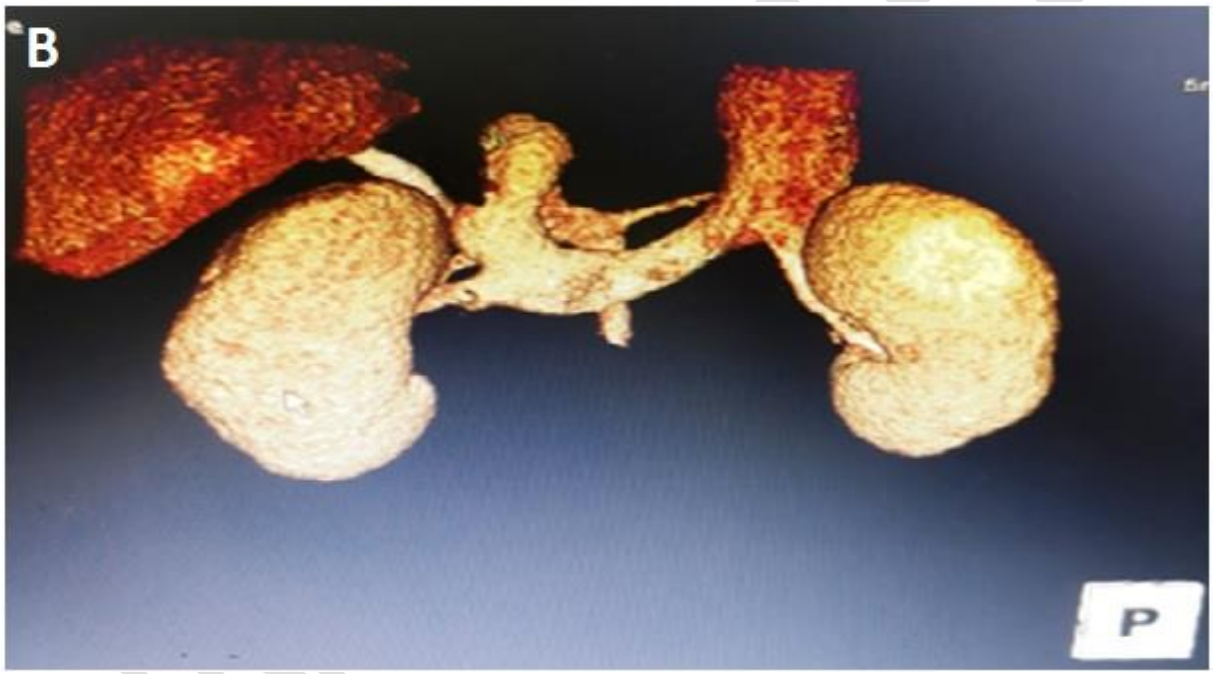
Figure A-B. CT axial sections showed the SMV and the splenic vein merges into the left renal vein drain. PV is thin-walled and hypoplastic.

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